

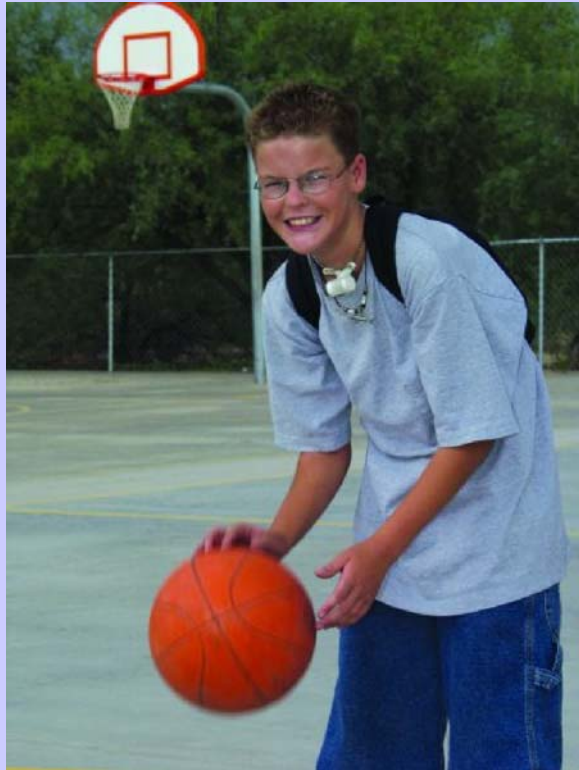
Southwest Ventilation Program, Caring for Children and Young Adults with Chronic Respiratory Problems

**Joanne Douthit, RN, MN
Clinical Nurse Specialist
Pediatric Pulmonary Section
Arizona Respiratory Center**



Learner Outcomes

1. Describe the goals and objectives of the Southwest Ventilation Program
2. Identify the role of family centered care to families that require assistive interventions to maintain normal breathing
3. Understand the need for education for patients, families and all health care providers in caring for this population with complex respiratory needs



Why?



Jimmy was born with a rare disorder called Congenital Central Hypoventilation Syndrome and requires ventilation assistance. By using implanted diaphragmatic pacers, Jimmy's breathing can be independent.



Need for Services



- Many pediatric and young adult patients will use complex respiratory therapies
- These individuals include debilitating lung diseases such as cystic fibrosis and neuromuscular problems
- No comprehensive ventilation center providing family centered care to patients and families exists in the region.
- Results in:
 - Fragmented care, multiple caregivers, multiple appointments
 - Crisis driven management
 - Lower quality of life
- Some families and patients travel thousands of miles to existing centers across the country to receive necessary treatment and equipment training

Resources

- **Conducted Community Needs Assessment**
- **Petitioned Community Durable Medical Equipment (DME) providers**
- **Financial assistance from Christopher Reeve Foundation**
- **Families as advocates**
 - **Philanthropic donations**
- **Pediatric Pulmonary Training Center**

Services Offered

Customized treatment and training from an experienced team using an inter- disciplinary approach

Access to state-of-the-art ventilation equipment and technology, available for training and patient/family

Regional education and care network built on existing infrastructure to deliver education, medical consultation, and direct care throughout Southwestern Arizona

Interdisciplinary Team

SVP's team will include:

- **Specially trained physicians**
- **Specially trained nurses**
- **Respiratory therapists**
- **Speech therapists**
- **Nutritionists and Social workers**
- **Physical therapists, occupational therapists**

The team will work with each patient and family, providing more comprehensive care and equipment training than could possibly be delivered into a short appointment with a single provider.



SVP Program Goals

- A. To improve care for people with complex ventilation needs*
- B. To implement effective education and training programs for patients, families, and medical professionals*
- C. To perform research, particularly clinical research, to improve the lives of people with complex respiratory needs*

Goals of the SVP



- **Establish protocols for the management of patients who require home ventilation and complex respiratory technology**
- **Provide individualized manuals of care to include anticipatory guidance and explanation of therapies and equipment**
- **Educational materials for healthcare professionals who care for these complex patients**

Cough Assist: Example



The Cough Assist Machine helps to clear secretions from the lungs by assisting you with your breathing. When you breathe in (inspiration), the machine gives you air (positive pressure) to help expand your lungs.

When you blow out (expiration), the machine creates a sucking force (negative pressure) that pulls the air out of your lungs. This rapid change in pressure during the different phases of breathing (inspiration and expiration) helps make your cough stronger and more effective.

Cough Assist cont.

Things to know about the Cough Assist Machine Cough Machine

The machine is very easy to learn once you understand what each knob controls.

- **Power Switch:** 'I' symbol means ON. 'O' symbol means OFF
- **Inhale:** determines how long the positive pressure will be blowing air into the lungs.
- **Exhale:** determines how long the negative pressure will be sucking or forcing the air out of the lungs.
- **Pause:** the time between the last negative pressure and next positive pressure.
- **Inhale Flow:** where you set the amount of flow needed during inspiration (taking a breath in). There are two settings: FULL or REDUCED. If REDUCED flow is used, there is a small reduction in inhale pressure.
- **Inhale Pressure:** you can adjust the inhale pressure to be less than exhale pressure.
- **Manual/Auto:** controls whether you want the machine to be automatically switching between pressures or if you want to do it by hand (manually).
- **Manual Control:** allows you to toggle between inhale and exhale when you

Post-Operative Care

RECOMMENDATIONS FOR POST-OPERATIVE CARE OF CHILDREN WITH NEUROMUSCULAR DISEASE FOLLOWING SURGERY (OR THE INTUBATED CHILD WITH NEUROMUSCULAR DISEASE)

For the intubated patient, perform the following q 4 hours:

Extubate when the patient is :

Following extubation perform the following q. 4 hours:

Wean from BiPAP during the day as tolerated. Goal is to use BiPAP per nasal mask while sleeping only. Wean airway clearance regimen above as airway secretions decrease working toward a four times during the day schedule or less.

Care of Children with Neuromuscular Disease during a Cold

During a cold, children with muscle weakness can have a very hard time clearing mucus from their lower airway. You can help them by:

- 1. These steps should be done in the order shown below every four hours and as needed while the child has a cold.**
 - 1. Chest physiotherapy for 10-20 minutes**
 - 2. Cough assist machine, four sets of 5 breaths**
 - 3. Postural drainage for 15-30 minutes**
 - 4. Cough assist machine, four sets of 5 breaths**
- 2. Use cough machine assist machine every time your child sounds rattley or has trouble coughing out secretions. If your child cannot get mucus out of the back of their throat, suction their mouth.**
- 3. If your child is on BiPAP, use the BiPAP machine with a nasal mask every time they are asleep. Also use it if your child is napping during the day. Your child is weaker than usual during colds.**

Respiratory Care for the Patient with Duchenne Muscular Dystrophy (DMD)

Evaluation and Anticipatory Guidance in DMD

- **DMD: X linked recessive, 1:3,000 male births, mutation of the dystrophin gene**
- **Diagnosis: Mutation analysis of blood leukocyte DNA of the dystrophin gene, confirmed by absent or abnormal dystrophin using immunohistology or protein analysis of muscle.**
- **Purpose: Educate practitioner about approaches and therapies available for the management of the respiratory complications of DMD. Many interventions listed may be applicable to other neuromuscular conditions. Patients with DMD should be evaluated routinely by providers trained in pulmonology, neurology, cardiology, nutrition, physical medicine, orthopedic surgery, mental health, sleep medicine, speech and social work.**

Routine Evaluation of Respiratory Function

History:

- 1. Cough (weak)**
- 2. Pneumonia**
- 3. Recurrent or persist bronchitis**
- 4. Asthma**
- 5. Sleep disturbances (frequent repositioning)**
- 6. Abnormal Chest radiograph**



Respiratory Evaluation

Pulmonary evaluation once a year if:

- **Between 4-6 years and/or not yet wheel chair bound for baseline pulmonary function testing, medical guidance regarding respiratory complications of DMD and assessment of ongoing respiratory therapies (if any)**

Pulmonary evaluations twice a year if:

- **Wheel chair bound**
- **Vital capacity less than 80% predicted**
- **Age 12 or older**

Pulmonary evaluation every 3-6 months if:

- **Requires mechanically assisted airway clearance therapy or mechanically ventilated**

Respiratory Evaluation

Pulmonary function testing

Awake Carbon dioxide tension

Other respiratory disorders to consider:

- Obstructive sleep apnea
- Oropharyngeal aspiration
- Gastroesophageal reflux
- Asthma

Sleep evaluation

- Review sleep quality and symptoms each visit
- Once wheelchair users, annual evaluation for sleep disordered breathing should be done or when clinically indicated

Management-DMD

- **Nutritional Issues**
- **Airway Clearance**
- **Manual Techniques**
- **Mechanical Techniques**
- **Mucus Mobilization Devices**
- **Speech Issues**

Management-DMD

- **Noninvasive Nocturnal Ventilation**
 - **BiPAP/CPAP**
 - **ventilators**
- **Daytime Noninvasive Ventilation**
 - **Mouthpiece intermittent positive pressure ventilation**
 - **Glossopharyngeal breathing**
 - **Intermittent abdominal pressure ventilator (Pneumo-belt)**
 - **Negative Pressure ventilation using chest cuirass**

Management-DMD

Continuous Invasive Ventilation

Tracheostomy:

- Advantages include more secure ventilator patient interface, ability to provide higher ventilator pressures if have intrinsic lung disease or severe reductions in chest wall compliance (eg scoliosis) and ability to perform direct airway suctioning
- Complications include generating more secretions, impairing swallowing, increasing risk of aspiration, bypassing airway defenses (increasing risk of infection), impairing oral communication, and acute airway obstruction (must be monitored to detect mucus plugs)

Education of Patient and Family

- A. Discussions about mechanical ventilation should be done before the need is apparent**
- B. Care of the technology-dependent young adult should be interdisciplinary**
- C. Every community should have a program for both children and adult including a transition process**

End of Life Care Goals

- A. Treating conditions (pain, dyspnea) that cause distress**
- B. Attending psychosocial and spiritual needs of the patient and families**
- C. Respecting the patient and family's choices concerning testing and treatments.**

Why?



Isabelle was born with a diaphragmatic hernia and undeveloped lungs that needed prolonged ventilation via a tracheostomy. As her lungs grow and develop, she will be weaned from mechanical ventilation.



Thanks!

“Be a good listener because you never learn much from talking.”

Will Rogers